Frequently Asked Questions
Classic Creutzfeldt-Jakob Disease (CJD)

What is Classic Creutzfeldt-Jakob Disease (CJD)?

- Classic Creutzfeldt-Jakob Disease (CJD) is a rare, fatal, degenerative brain disease caused by abnormal, transmissible proteins called prions.
- There are three types of classic CJD:
  - Sporadic CJD (occurs occasionally with no known cause)
  - Familial CJD (an inherited form of CJD that occurs in families)
  - Iatrogenic CJD (occurs in a patient who was infected during a medical or surgical procedure)

Further information about these specific types of classic CJD is included below.

How common is classic CJD?

- Classic CJD occurs in about 1 person per million people each year.
- Between 0 and 6 cases of CJD each year would be expected in Utah residents. Actual detected cases of CJD in Utah have been within the expected numbers.

Year	Number of cases of CJD reported in Utah
2002	2
2003	1
2004	0
2005 (as of November 16, 2005)	0

  - Sporadic CJD accounts for 85-90% of diagnosed classic CJD cases.
  - Familial CJD accounts for 10-15% of diagnosed classic CJD cases.
  - Iatrogenic CJD accounts for <1% of diagnosed classic CJD cases.

What are the symptoms of classic CJD?

- Initial symptoms vary from patient to patient. Symptoms can include mental deterioration or dementia, memory loss, difficulty with balance and walking, dizziness, behavioral changes, visual disturbances, and involuntary movements (called myoclonus).
- In the advanced stages of the disease, patients commonly develop significant difficulties in movement and become unable to talk and swallow.

How is classic CJD diagnosed?

- After symptoms appear, doctors often can make a preliminary diagnosis based on medical history, physical examination of the neurological system, and certain diagnostic tests including:
  - Electroencephalogram (EEG)
  - Magnetic resonance imaging (MRI)
  - Spinal fluid tests
- A confirmed diagnosis of CJD can usually only be made after the patient's death.

How is classic CJD treated?

- Symptoms of the disease are treated, but there is no treatment available that slows or stops the disease.

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Is there a vaccine or medicine available that cures or prevents classic CJD?

- No vaccine is currently available to prevent classic CJD, and no medicines have been identified to cure or prevent the disease.

What is being done to prevent classic CJD?

- Research is being done to better determine what causes CJD and identify possible treatments for the disease.

Is classic CJD reportable in Utah?

- Both classic CJD and vCJD should be reported to your local health department or the Utah Department of Health.

SPORADIC CREUTZFELDT-JAKOB DISEASE (CJD)

What is Sporadic Creutzfeldt-Jakob Disease (CJD)?

- Sporadic CJD is a very rare, fatal, degenerative brain disease that occurs spontaneously in the general population with no known cause or triggering event.

What are the symptoms of sporadic CJD?

- In typical cases of sporadic CJD, early symptoms often include depression, mood swings, memory lapses, social withdrawal, and lack of interest. Rapid progression to dementia and obvious neurological symptoms distinguish sporadic CJD from clinical depression.
    - Within weeks of diagnosis, the patient may become unsteady on his/her feet, lacking in coordination and becoming markedly clumsy. In some people, these are the first symptoms of disease. This pattern of symptoms is known clinically as cerebellar ataxia because it is caused by damage to the cerebellum, the part of the brain that controls movement.
    - Later symptoms may include blurred vision, hallucinations, blindness, rigidity in the limbs, sudden jerky movements and incontinence.
    - Speech may become more difficult or slurred. Swallowing may become difficult.
    - Eventually, the patient loses the ability to move or speak and will require full time nursing care. In this state (clinically known as akinetic mutism)- the patient may appear to be following what is going on around them, but in fact they are not aware of their surroundings.
    - Most patients die within a few months of onset of symptoms.

What are the risk factors for sporadic CJD?

- There are no known specific risk factors for sporadic CJD.
- Most cases are identified in individuals between 45 and 75 years of age; the median age at death for diagnosed cases is 68 years.

What causes sporadic CJD?

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The cause of sporadic CJD is not known. Several studies have not found evidence that it is acquired as an infection. Currently, it is generally believed that it occurs through a spontaneous mutation leading to production of the abnormal protein, or prion.

FAMILIAL CREUTZFELDT-JAKOB DISEASE (CJD)

What is Familial Creutzfeldt-Jakob Disease (CJD)?

- Familial Creutzfeldt-Jakob Disease (CJD) is an uncommon inherited form of CJD.

What are the symptoms of familial CJD?

- The symptoms of the familial form of CJD vary, depending on the type of mutation involved. There may even be variation in symptoms within affected members of the same family.
- Symptoms may include but are not limited to:
  - Depression, bizarre or uncharacteristic behavior, and memory lapses may be evident.
  - Fatigue and visual disturbances may be reported.
  - Within weeks of onset of symptoms, unsteadiness (gait ataxia) and lack of coordination (cerebellar ataxia) may occur.
  - Difficulties with speech and/or swallowing may develop.
  - Sudden jerky movements (myoclonus), rigid limbs, blindness and incontinence may also be associated with disease.

Who can get familial CJD?

- Clusters of familial CJD are found in families because in this form of disease, people inherit abnormal proteins, making them more prone to developing disease.
- Often familial CJD is diagnosed at an earlier age than is typical for other forms of CJD. For example, the average age of onset is approximately 52 years compared to 60 years for cases of sporadic CJD.

How is familial CJD diagnosed?

- In addition to a medical history, physical examination, and testing used to diagnosis the sporadic form of classic CJD, a family history and genetic testing are used to determine if the cause is inherited or familial as opposed to being sporadic.

IATROGENIC CREUTZFELDT-JAKOB DISEASE (CJD)

What is Iatrogenic Creutzfeldt-Jakob Disease (CJD)?

- Iatrogenic Creutzfeldt-Jakob Disease (CJD) is CJD acquired through a medical or surgical treatment or a diagnostic procedure.

What are the symptoms of iatrogenic CJD?

- When exposure to infected materials occurs during an invasive procedure of the brain, symptoms are like those of sporadic CJD. However, when iatrogenic CJD is acquired through invasive procedures not directly affecting the brain, symptoms usually include lack of

coordination and unsteadiness (ataxia); in this case, dementia may not occur until late in the course of the disease.

How is iatrogenic CJD transmitted?

- Iatrogenic CJD has been documented to occur in the following circumstances:
  - Persons receiving injections of pituitary hormones from infected cadavers.
  - Persons receiving corneal transplants from infected donors.
  - Persons exposed to contaminated surgical instruments used on the brain during surgery.
  - Persons exposed to contaminated electrodes used during electroencephalographic (EEG) procedures.
  - Persons receiving dura mater grafts from infected donors.
  - Persons receiving a transfusion of blood or blood product from a person with variant CJD.
- Transmission of CJD directly from one person to another person, other than by the means listed above, has not been documented.

What is the incubation period for iatrogenic CJD?

- The incubation period (time from infection until onset of illness) for iatrogenic CJD can be as short as 1 year to 30 years or longer. It varies depending on the way the infection was transmitted; incubation period tends to be longer for exposures that occur through the blood stream (such as with pituitary-derived hormones) than for those where the exposure involves the central nervous system (such as with exposure to a contaminated instrument or dura mater graft).

What is done to prevent the spread of iatrogenic CJD?

- Hospitals have strict guidelines to sterilize neurosurgery instruments.
- Instruments used on the brain of someone with suspected CJD are destroyed.
- Since 1985, growth hormone has been made synthetically rather than being extracted from human pituitary glands, so there is no current risk from this source.
- Since 1992 human dura mater grafts have not been used; human tissue has been replaced with a synthetic substitute.
- There is no alternative to using corneas taken from human donors after death for cornea transplants. However, risk associated with this procedure appears to be minimal, as many thousands of grafts are performed annually but only 2-3 cases of corneal graft-related CJD have ever been recorded worldwide. Also, organ donation is very tightly controlled. Transplants are not taken from donors diagnosed with CJD. In fact, organ and tissue donations of any kind are not accepted from suspect cases of CJD (classic or variant) in order to limit the possibility of transmission of the disease.

Where can people get more information?

- More information is available from your physician, the Creutzfeldt-Jakob Disease Foundation (330-665-5590) www.cjdfoundation.org, the CJD Surveillance National Prion Disease Pathology Surveillance Center (216-368-0587) www.cjdsurveillance.com, your local health department, the Utah Department of Health (801-538-6191), or the Centers for Disease Control and Prevention (CDC) http://www.cdc.gov/ncidod/dvrd/cjd/ .